

Early Diagnosis Of Rheumatic Fever

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Introduction:

Rheumatic fever causes chronic progressive damage to the heart and its valves. Until 1960, it was a leading cause of death in children and a common cause of structural heart disease. The disease has been known for many centuries. Baillou (1538-1616) first distinguished acute arthritis from gout. Sydenham (1624-1668) described chorea but did not associate it with acute rheumatic fever (ARF). In 1812, Charles Wells associated rheumatism with carditis and provided the first description of the subcutaneous nodules. In 1836, Jean-Baptiste Bouillaud and, in 1889, Walter Cheadle published classic works on the subject

The association between sore throat and rheumatic fever was not made until 1880. The connection with scarlet fever was made in the early 1900s. In 1944, the Jones criteria were formulated to assist disease identification. These criteria, with some modification, remain in use today. The introduction of antibiotics in the late 1940s allowed for the development of treatment and preventive strategies. The dramatic decline in the incidence of rheumatic fever is thought to be largely owing to antibiotic treatment of streptococcal infection.

Pathophysiology:

ARF is a sequel of a previous groups A streptococcal infection, usually of the upper respiratory tract. One beta-streptococcal serotype (e.g., M types 3, 5, 18, 19, 24) is linked directly to ARF. The disease involves the heart, joints, central nervous system (CNS), skin, and subcutaneous tissues. It is characterized by an exudative and proliferative inflammatory lesion of the connective tissues, especially that of the heart, joints, blood vessels, and subcutaneous tissue.

Mortality/Morbidity:

Morbidity from ARF is directly proportional to the rate of streptococcal infections. Infections that are not treated adequately are most likely to cause the major sequelae noted in the list of Jones criteria. Morbidity is also related to the care that the patient receives.

The mortality rate has declined steadily over the last 3 decades. A partial explanation for the decrease in mortality rate may be the increase in antibiotic use. In developing nations like ours with majority of lower socioeconomic areas where rheumatic fever is more prevalent, ARF is a major cause of death and disability in children and adolescents.

Cardiac involvement is the major cause of long-term morbidity. Migratory polyarthritis occurs early in the disease course and is a common complaint for patients with rheumatic fever. Joint involvement ranges from arthralgia without objective findings to overt arthritis with warmth, swelling, redness, and exquisite

tenderness. The larger joints are involved most frequently, such as the knees, ankles, elbows, and wrists. An inverse relationship between severity of joint involvement and risk of carditis appears to exist.

- In approximately 75% of cases, the acute attack lasts only 6 weeks.
- Ninety percent of cases resolve in 12 weeks or less.
- Fewer than 5% of patients have symptoms that persist for 6 months or more.

Sex:

No sex predilection exists, except that mitral valve prolapse and Sydenham chorea occur more often in females than in males. For our subcontinent it seems males are affected more than females.

Age:

Although individuals of any age group may be affected, most cases are reported in persons aged 5-15 years. For our subcontinent it is below 19 years of age.

CLINICAL PRESENTATION:

Case 1: A 21 years old male was referred with complaints of fever on & off for 15 days and pain of bilateral knee joints and bilateral elbow joints for 15 days. He was asymptomatic 1 month back when he developed Sore throat followed by fever which was on and off, low grade, not associated with chills and rigor, fever showed no periodicity. Pain in both knee joints started 2 weeks later, both elbow joints followed next which got so severe to the extent that he could not eat and walk. He gave h/o mild non-tender nodule in the right upper shin 3-4 days after onset of joint pain but it has subsided now.

Case 2: A 23 years old female presented with only pain in the left ankle joint. She went to orthopedic surgeons and innumerable general physicians but with no relief of symptoms. When she came to us she gave history of multiple episodes of sore throat. On auscultation patient had mild MR. On subjecting the patient to 2D-Echo she had valvulitis of the mitral valve. Finally she was diagnosed as a case of rheumatic fever.

Case 3: A 14 year old presented with fever on & off for 15 days, & pain in bilateral knee joints for 15 days. He was treated outside as a case of enteric fever with no improvement. When he was referred to us he had a small subcutaneous nodule on his right upper shin from which we could diagnose him as a case of rheumatic fever.

Case 4: A 19 years old boy presented with fever of low grade for 2 weeks duration with pain in all big joints. His physical examination revealed no abnormalities. On subjecting the patient to 2D-Echo he was found to have valvulitis of mitral valve.

WORKUP

Lab Studies:

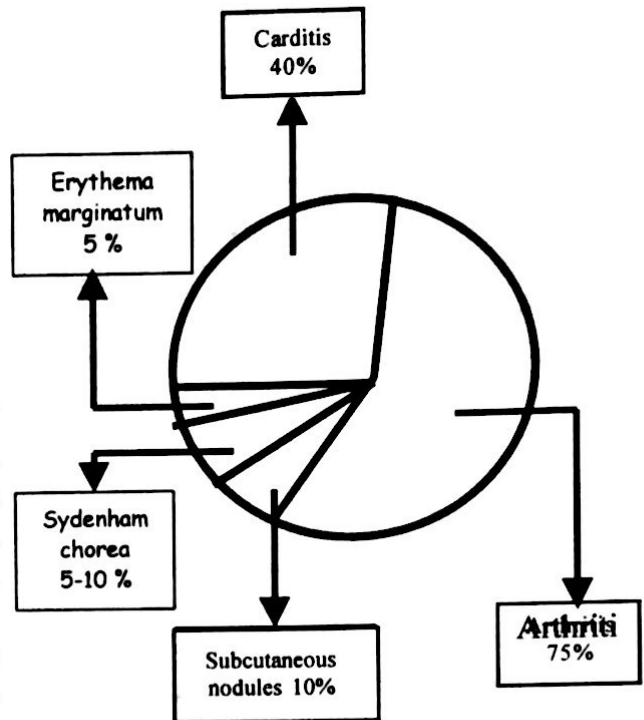
1. Blood Routine :
T.L.C: raised.
D.L.C: leucocytosis may be present.
E.S.R: raised.
Hb: reduced.
2. Rheumatoid Factor: positive.
3. A.S.O titre: positive >200 U/L.
4. C.R.P: positive.
5. Chest X-ray- features suggestive of cardiomegaly or CCF.
6. ECG – variable depending upon the type of presentation.
7. S.Uric Acid- to rule out any joint pathology.

8. Blood from 3 different sites for C/S : to rule out I.Endocarditis
9. Throat swab for C/S : suggestive of sorethroat.
10. Echocardiography may show AML, PML thickening, valvulitis with or without murmurs suggestive of rheumatic aetiology.

Diagnosis: The diagnosis requires 2 major Jones criteria or 1 major plus 2 minor Jones criteria.

Major criteria

1. Carditis occurs in as many as 40% of patients and may include cardiomegaly, new murmur, congestive heart failure, and pericarditis, with or without a rub and valvular disease.
2. Arthritis occurs in 75% of cases and is polyarticular, fleeting, and involves large joints.
3. Subcutaneous nodules (ie, Aschoff bodies) occur in 10% of patients and are edematous fragmented collagen fibers. They are firm painless nodules on the extensor surfaces of wrists, elbows, and knees.
4. Erythema marginatum occurs in about 5% of cases. The rash is serpiginous and long lasting.
5. Sydenham chorea (ie, St. Vitus dance) is a characteristic movement disorder that occurs in 5-10% of cases. Sydenham chorea consists of rapid purposeless movements of the face and upper extremities. Onset may be delayed for several months and may cease when the patient is asleep.



Minor criteria

1. Clinical findings include arthralgia and fever. Both was present in our case under study.
2. Laboratory findings include elevated acute phase reactants {eg, erythrocyte sedimentation rate (E.S.R), C reactive protein (C.R.P)}, & a prolonged PR interval.

Supporting Evidence

1. Antecedent group A streptococcal infections (i.e, positive throat culture or rapid streptococcal screen and an elevated or rising streptococcal antibody titer).
2. Recent scarlet fever.
3. Increased streptococcal antibodies.

Complications:

1. Congestive cardiac failure.
2. Rheumatic valvular heart disease. Eg. Mitral stenosis, mitral regurgitation, aortic stenosis, aortic regurgitation,
3. Arrhythmias and thrombo-embolic events.
4. Pericarditis with effusion.
5. Rheumatic peritonitis.

TREATMENT

Departmental Care:

Our responsibilities were to suspect the diagnosis and to treat complications. We considered early administration of antibiotics.

Consultations:

Because of the many clinical features of ARF, consulting a cardiologist, a rheumatologist, and a neurologist is necessary. Carditis is not only a major clinical finding, but it also is the cause of much of the disability. Arthritis is one of the major manifestations. Movement disorders associated with ARF may be difficult to differentiate from those of other clinical problems.

Medication:

Medical therapy involves the following 5 areas:

1. Treatment of group A streptococcal infection was considered regardless of organism detected.
2. Steroids and salicylates were used to control pain and inflammation.
3. If heart failure is present than digitalis can be administered.
4. Administer prophylaxis to patients who have developed ARF. The most important way to prevent rheumatic fever is by proper and prompt treatment of streptococcal throat and scarlet fever.
5. Haloperidol may be helpful in controlling chorea.

Our case under study was given following treatment:

1. **Bed rest:**
Our patients were advised bed rest till temperature and E.S.R were normal.
2. **Antistreptococcal therapy:**
A course of antibiotic was given to eradicate the streptococci even if throat culture was negative. One of the following regimen can be used:
 - a) Single inj. of benzathine penicillin 1.2 MU i.m.
 - b) Dailly inj. of procaine penicillin 600.000 units i.m for 10 days.
 - c) Oral Erythromycin 20-40 mg/kg/day in four divided doses (in those cases sensitive to penicillins).
3. **Salicylates:**
Aspirin at doses of 60 mg/kg/day in 6 divided doses. Can be increased upto 120mg/kg/day or a maximum of 8 gm per day. Aspirin should be continued till E.S.R is normal and then gradually tapered off over 4-6 weeks.
4. **Corticosteroids:**
Prednisolone at 60-120 mg/day in four divided doses till ESR is normal and then gradually tapered off over a period of 2 weeks.

Indications:

- a. In severe carditis manifested by CCF not responding to aspirin.
- b. In severe arthritis not relieved by aspirin.
5. **Supportive therapy :**
Includes treatment of CCF, valvular lesions, heart blocks and chorea.
6. **Prevention of rheumatic fever:**
 - a. **Primary Prevention:**
Accurate diagnosis and treatment of group A streptococcal pharyngitis infection. Established streptococcal pharyngitis can be treated by benzathine penicillin or oral penicillin or erythromycin.
 - b. **Secondary Prevention (Rheumatic Fever Prophylaxis):**
Should be given in all cases who has a documented attack of rheumatic fever.

Regimens:

- # Three monthly i.m. inj. of 1.2 MU of Benzathine Penicillin (most effective).
- # Oral Penicillin V 250 mg b.i.d.

Tab. Sulphadiazine 1 gm/day orally as a single dose (in those allergic to penicillins)

Tab. Erythromycin 250 mg b.i.d. (in those allergic to penicillins).

Durations :

- Upto the age of 25 years.
- If after 25 years of age then till 5 years after last exposure.
- If second attack of rheumatic fever or with R.H.D then till the age of 40 years or 10 years after the second episode.

DISCUSSION:

Disease prevalence here in Nepal & other countries of this subcontinent depends largely upon socioeconomic status, with higher frequency in areas of crowding. Because of varied atypical presentation and lack of diagnostic investigations in all the centers it is often missed in our subcontinent, though quite a lot of cases persist in our community. Frequency of streptococcal infection and virulence of the bacterial strain determine the incidence of rheumatic fever in the population. Diagnosis requires a high index of clinical suspicion.

As a sequel of beta-streptococcal exposure, ARF occurs during school years when streptococcal pharyngitis is most prevalent. Similarly, prevalence is higher in the colder months of the year when streptococcal pharyngitis is most likely to occur. Onset of rheumatic fever to rheumatic heart disease is 1-2 years. Only 50% gives h/o rheumatic fever. Thrombo-embolism, calcification and atrial fibrillation are less common. The rate of development of acute rheumatic fever in individuals with untreated streptococcal infection is estimated to be 3%. Persons who have suffered from rheumatic fever have a tendency to develop flare-ups with repeated streptococcal infections. Although rare, acute rheumatic fever can cause heart failure and death. It most often requires hospitalization. Because of treatment with antibiotics, recurrence of the disease is rare. However, the risk of valvular heart disease is increased. Following treatment, frequent checkups to monitor valve damage may be necessary. Internationally ARF has increased incidence in developing countries. As many as 25-40% of cases worldwide appear in third world nations. For complicated cases here in Nepal, the procedure of Closed Mitral Valvotomy was introduced in Bir Hospital during late 1980's. Closed Mitral Valvotomy was successfully started in Shree Birendra Mititary Hospital in the year 1995.

Conclusion:

Finally it is important to say that rheumatic fever has varied presentation and can be missed easily during day to day practice, so a thorough history, clinical examination and high index of suspicion supported by the laboratory and Echocardiography findings are necessary to diagnose the case of rheumatic fever early. This can also propagate the information regarding preventive cardiology.

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